Researchers test the systemic score for Still's disease

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Still's disease is an inflammatory disorder typically manifesting with fever, arthritis, and skin rash. It affects both children and adults, and has previously been known as systemic juvenile idiopathic arthritis and adult-

onset Still's disease. Regardless of the name, people with Still's disease are burdened by life-threatening complications.

A multi-center, observational, prospective study was designed to evaluate the clinical usefulness of the systemic score in predicting life-threatening evolution—defined as the development of macrophage activation syndrome (MAS) and/or mortality. The intention was also to derive a more aggressive clinical patient subset.

To achieve this, Ruscitti and colleagues collected data from 597 patients taking part in the GIRRCS (Gruppo Italiano Di Ricerca in Reumatologia Clinica e Sperimentale) AOSD-study group and in the AIDA (AutoInflammatory Disease Alliance) Still's disease registry. First developed in the 1990s, the prognostic value of the systemic score has previously been tested in a small number of cases. The new work was presented in a session on the management of Still's disease across the life-span at the 2024 congress of EULAR—The European Alliance of Rheumatology Associations.

Each patient was assessed for the systemic score, which assigns 1 point to each of 12 manifestations: fever, typical rash, pleuritis, pneumonia, pericarditis, hepatomegaly or abnormal liver function tests, splenomegaly, lymphadenopathy, leucocytosis > 15,000/mm^3, sore throat, myalgia, and abdominal pain.

When looking at manifestations, 100% of patients included had fever, 87.9% had joint involvement, and 66.1% had skin rash. Liver involvement was recognized in 43.5%, MAS in 13.1%, and lung disease in 6.9%. There was a mortality rate of 3.4% attributed to Still's disease.

A result of at least 7 on the systemic score proved to be a significant predictor of life-threatening evolution. To further explore this, a risk-profile assessment was performed on the clinical variables used to

calculate the systemic score, and an age- and sex-adjusted multivariate logistic regression model built. This showed that liver involvement and lung disease independently predicted life-threatening evolution.

The clinical characteristics of this subset of patients was then derived, revealing that those with liver involvement were significantly characterized by lymph-adenomegaly, splenomegaly, pericarditis, and pleuritis—and were also burdened by lung disease.

In the patients with lung disease, significant characteristics were sore throat, lymph-adenomegaly, splenomegaly, and liver involvement. Furthermore, those with lung disease showed a higher frequency of pericarditis, pleuritis, and abdominal pain. This subset was also burdened with a higher mortality rate.

The results suggest that the systemic score could be used as a prognostic tool in clinical practice. Additionally, liver involvement and lung disease are clinically relevant findings, and should be highlighted as key multi-organ manifestations of Still's disease, and as major predictors of life-threatening evolution.

More information: P. Ruscitti et al, OP0001 The evaluation of systemic score in identifying patients with Still's disease at higher risk of life-threatening evolution; findings from GIRRCS AOSD-study group and AIDA Network Still's Disease Registry, *Scientific Abstracts* (2024). DOI: 10.1136/annrheumdis-2024-eular.667

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